

be performed with urine organic acids so that this diagnosis can be quickly made. With the emergence of new genetic therapies for biochemical diseases it may be important in the future to make this diagnosis early in the course of the disease.

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Transient myasthenia gravis in an elderly woman

Myasthenia gravis is usually a chronic disorder, although remission rates of 25-30% can be expected with judicious interventions such as thymectomy and immunosuppression in appropriate cases. Spontaneous remission occurs rarely. Oosterhuis found a remission rate of only 1% per annum over 17 years among 180 patients with generalised myasthenia gravis treated with anticholinesterases alone.¹ We describe a case of ocular and bulbar myasthenia in which there was complete recovery over a matter of weeks without immunosuppressive agents.

The patient, a 78 year old woman, developed acute, painless left sided ptosis in October 2002, while on holiday abroad. There was no diplopia. She had developed hypothyroidism one year before but was euthyroid on replacement therapy. The ptosis resolved gradually over the next few days. There was no family history of autoimmune diseases. Two weeks later she found she could not chew properly towards the end of a meal. The jaw weakness recovered the following day but later recurred, to the extent that she had to support her jaw while talking and eating. She had slight dysphagia but there was no history of choking, breathing problems, or limb weakness.

On examination in January 2003, there was no definite ptosis or ophthalmoparesis. No fatigability was demonstrated in the eyelids, and no weakness or fatigability was

detected on testing jaw opening against resistance, although her jaw hung slackly open if not supported. Limb power and reflexes were normal.

Acetylcholine receptor antibodies (AChR-*abs*) were strongly positive at 11 nmol/l (normal range 0 to 0.25 nmol/l). Other autoantibodies were negative apart from a positive gastric parietal cell antibody. Computed tomography of the mediastinum did not show any thymic abnormality. She was given pyridostigmine 60 mg twice daily, and her symptoms gradually improved. On repeat assessment one month later, there was no evidence of ptosis and the jaw weakness seemed to be improving, though she still needed to hold her jaw when speaking for a few minutes. Repetitive nerve stimulation of left frontalis and obicularis oculi showed no decremental response. Electromyographic examination of the right biceps, extensor digitorum communis, and left frontalis muscles was also normal.

In view of the improving symptoms and the electrophysiological findings, it was decided to continue pyridostigmine alone and steroid treatment was not initiated. Her symptoms continued to improve. By April 2003, she was completely asymptomatic. The pyridostigmine was gradually withdrawn over one month. She remained asymptomatic on follow up in September 2003. Neurological examination was entirely normal although her AChR-*abs* remained positive at 5.00 nmol/l.

COMMENT

To our knowledge, transient myasthenia gravis has not been described previously in a patient of this age. It is known to occur in 10-15% of infants born to mothers with myasthenia as a result of transplacental transfer of maternal antibodies, and there is a report of transient myasthenia in autoimmune disease resulting from HIV infection.² In that study, seven HIV infected patients presented with transient myasthenic symptoms. Four of them had positive AChR-*abs*. In one, myasthenia gravis and coincident autoimmune thrombocytopenia both resolved following anti-retroviral treatment. Our patient had no history of preceding infection, although there was a history of thyroid disease.

The persistence of high levels of AChR-*abs* despite clinical resolution is of interest. There is no direct correlation between antibody titre and clinical state in individual myasthenic patients.³ The resolution of clinical symptoms and signs, despite persisting acetylcholine receptor antibodies, presumably reflects a positive balance of acetylcholine receptor genesis to antibody mediated destruction.

Late onset myasthenia gravis differs from the typical form in several ways. The HLA DR3 haplotype is uncommon in late onset disease, while thymomas are more common. It is said that late onset disease is more severe and less likely to remit, and that bulbar involvement is more common.⁴ Response to conventional treatment is also less satisfactory than in younger patients. Our patient was therefore atypical of this older group.

A recent cohort study suggested that myasthenia gravis might be underdiagnosed in older people.⁵ In that study, 2000 asymptomatic individuals aged 60 and over, who participated in the Oxford healthy aging project, were screened for AChR-*abs*. Surprisingly, 0.71% were seropositive. Four

of eight seropositive subjects had a diagnosis of "stroke" or "transient ischaemic attacks", and the authors concluded that these patients might have been misdiagnosed.

It is therefore possible that myasthenia gravis in the elderly is sometimes missed and that the clinical course may in some cases be more benign than appreciated.

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"Doctor, I can hear my eyes": report of two cases with different mechanisms

An unusual but fascinating symptom is the one described by patients complaining that they can hear their own eye movements. We report two cases, with different postulated mechanisms.

Case reports

Patient 1

A 53 year old woman presented with a tendency to fall to the left. She did not experience hearing difficulties or tinnitus. Clinical examination was unremarkable other than increased sway on Romberg. Pure tone audiometry was normal other than bilaterally mildly elevated thresholds of 30 dBHL at 8 kHz. However left sided ipsi- and contra-lateral stapedial reflexes were elevated, and auditory brain-stem evoked responses, while normal on the right, showed increased latency of wave V on the left. Serial MRI scans demonstrated an enlarging left vestibular schwannoma. Trans-labyrinthine resection of the tumour was complicated by a left cerebellar infarct and hydrocephalus, and subsequently the patient continued to experience imbalance. Two years post-operatively, direct questioning about tinnitus led to the patient describing tonal tinnitus in the left ear on looking to the left, in which up-gaze increased the pitch, while down-gaze lowered the pitch. Notably she said "I feel I could play a tune with my eyes".

Patient 2

A 32 year old man presented with visual instability and a tendency to fall forward and to the left provoked by loud sounds such as

the telephone ringing in his left ear. His auditory symptoms included being able to hear his heart beats, bone taps, and footsteps. Additionally he complained of a soft low pitched sound in his left ear "rather like moving a hard-pressed finger across a clean, wet china dinner plate" when he moved his eyes. These symptoms could be reduced if the patient tensed his abdominal muscles. Pure tone audiometry showed normal air conduction hearing thresholds bilaterally other than a mild elevation of 35 dBHL at 0.25 kHz on the left. Bone conduction hearing levels were normal, perhaps even supra-normal, with thresholds of -10 dBHL at 0.5 and 1 kHz bilaterally, giving rise to an "air-bone gap" at these frequencies. Clinical examination and three-dimensional video oculography demonstrated left beating torsional nystagmus provoked by the patient humming, and CT scanning of the petrous temporal bones revealed bilateral dehiscence of the superior semicircular canals (fig 1).

Discussion

Patient 1 has gaze-evoked tinnitus, a phenomenon first described in 1982¹ that was initially thought to be rare, but subsequently reported to be surprisingly common (prevalence 19–36%) in one study of patients post vestibular schwannoma resection.² It has also been described in patients with cerebello-pontine angle meningioma, meningeal metastases of malignant melanoma, and sudden sensorineural hearing loss.² It may develop months post-operatively, and is usually heard in, and caused by moving the eyes towards, the diseased ear. The exact mechanism is not known, but it has been postulated that neural plasticity mechanisms activated by unilateral deafferentation result in cross-talk between neural elements controlling eye movements and the central auditory system. Indeed, functional imaging studies of patients with gaze-evoked tinnitus have shown anomalous activation of the auditory lateral pons and auditory cortex, enhanced by failure of cross-modality inhibitory mechanisms.³

Patient 2 has Tullio phenomenon, a condition in which sound and/or pressure stimulates the vestibular system. These patients often complain of abnormal auditory sensations such as "hearing footsteps vibrating through the body", and finding the noise of

chewing loud enough to make understanding of conversations at mealtimes difficult. Supra-normal bone conduction thresholds, or so-called "conductive hyperacusis", has been reported in other patients with Tullio phenomenon,⁴ and dehiscence of the superior semi-circular canals is thought to be the commonest associated pathology.⁵ It is postulated that the dehiscence may act as an alternative lower impedance pathway for sound energy, enabling these patients to hear the movements of their eyeballs within the bony sockets, these sounds being conducted through the skull. It is interesting to note that although our patient had dehiscence of both superior semi-circular canals, he was only symptomatic on the left side. This could be explained by the Tullio phenomenon being multi-factorial in nature, and additional factors such as trauma or bone remodelling need to be present as well as the dehiscence before the clinical features become manifest.⁵

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Transient compulsive hyperphagia in a patient with a thalamic infarct

Eating disorders are associated with various psychiatric and neurological diseases. Pathological eating behaviour ranges from reduced to excessive appetite, dysregulation of hunger and satiation signals, and odd food preferences. Hyperphagia and anorexia are reported in lesions (mostly tumours) involving the ventromedial hypothalamus.¹ Moreover, eating disorders also occur in temporal lobe tumours, temporal lobe epilepsy, and advanced states of degenerative disease with neuronal loss in the medial temporal lobe. Hyperorality is part of the Kluver-Bucy syndrome which occurs in patients with bilateral mesial temporal lesions.¹

We report a patient in whom compulsive hyperphagia was associated with a medial thalamic ischaemic stroke.

CASE REPORT

A 52 year old man complained of diplopia, dizziness, vertigo, decrease of consciousness, memory impairment, and hyperphagia. These symptoms occurred abruptly while he was painting the gate in his garden.

The diplopia and vertigo disappeared in about 15 minutes, while hyperphagia and memory impairment lasted for about 24 hours and completely disappeared the following day on awakening. Instead, amnesia concerning the event is still present.

The decrease of consciousness consisted of slight hypersomnolence (the patient could easily be awakened by auditory and verbal stimulation) and was present only at the onset, while memory impairment was noted only later. The patient's wife reported that he kept repeating the same questions to her, and painted the garden gate incorrectly (using colours inappropriately). The patient was completely unaware of these symptoms, which were reported only by his wife.

Regarding the hyperphagia, his wife reported that the patient compulsively ate all the food he found in the refrigerator and in the kitchen. Apart from the fact that he was eating continuously all day long, when it was time for his lunch and evening meal, he always felt hungry and ready to eat, as his main concern was food, and his attention could not be diverted during his meals.

The following day his wife took him to the hospital. Neurological examination on admission revealed only retrograde amnesia about the events that had occurred in the previous 24 hours, along with fatuous behaviour. The medical history was not significant. Cranial computed tomography and extracranial Duplex ultrasonography were normal. Five days later, brain magnetic resonance imaging (MRI) revealed an ischaemic lesion involving the medial portion of the left thalamus in the territory of the tubero-thalamic perforating artery (fig 1). MRI-angiography of extracranial and intracranial cerebral arteries was normal.

A transthoracic echocardiogram was normal, while a transoesophageal echocardiogram revealed an atrial septal aneurysm without a patent foramen ovale. Conventional vascular risk factors (hypertension, diabetes, hypercholesterolaemia, and smoking) were absent. Routine laboratory studies including coagulation profiles were normal.

During the hospital admission, the patient's clinical condition progressively improved and he was discharged eight days later without any neurological symptoms.

Two weeks later he was submitted to a neuropsychological test battery to assess reasoning (Raven PM47), short term memory (verbal and spatial span), learning (paired associate and Corsi spatial learning), attention (visual search), and frontal executive functions (Nelson card sorting test, trial making B, category, and literal fluency). Anxiety and depression were investigated by the state-trait anxiety inventory and the Beck depression inventory. Results were within the normal range (<1 SD) for all the tests.

COMMENT

The anatomical substrates of eating disorders are conventionally the basal ganglia and the cortico-limbic areas. In our patient compulsive hyperphagia was associated with the clinical features of medial thalamic ischaemia. The patient presented with signs and symptoms of the so called "top of the basilar

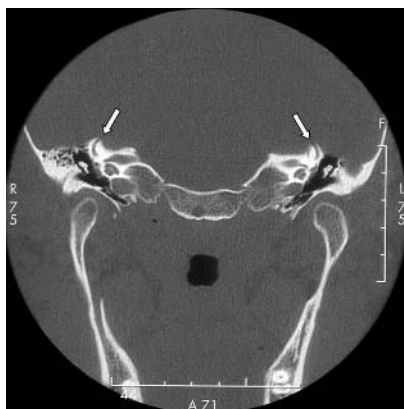


Figure 1 Transverse computerised tomography image showing bilateral dehiscence of the superior semi-circular canals (indicated by arrows).